

Congenital cardiac rhabdomyomas in red wattle pigs

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Rhabdomyomas are relatively rare, primary tumors of the cardiac muscle that occur sporadically in pigs, sheep, cattle, and dogs (1,2). Primarily observed at slaughter, or in neonates that have died due to other causes, cardiac rhabdomyomas are usually deemed to be incidental lesions in domestic animals (1). To date, there is no indication of a familial predisposition for cardiac rhabdomyomas in domestic animals. In this paper, multiple cardiac rhabdomyomas in stillborn and neonatal red wattle and red wattle-cross piglets are described, which is the first report that confirms a breed predisposition of these tumors in domestic animals. According to the Red Wattle Hog Association of Canada, domestic red wattle pigs have descended from feral red wattle pigs in Texas and are named for their color and the distinctive appendages that hang from their jawline (3).

From April 1988 to September 1992, of the 5908 total porcine accessions to all laboratories in the Veterinary Laboratory Services Branch (VLSB), Ontario Ministry of Agriculture and Food, only five were red wattle or red wattle-cross pigs. Information regarding the other breed(s) within each cross was not available. Cardiac rhabdomyomas were diagnosed in three of these consignments, one of which consisted of multiple red wattle-cross piglets. This group was composed of three, live, five-day-old piglets with diarrhea that were subsequently confirmed with coronavirus atrophic enteritis, and four stillborn term piglets retrieved from a sow that was acutely ill with transmissible gastroenteritis. The other submissions consisted of a 10-week-old red wattle-cross pig with severe bacterial myositis and septicemia, and a two-week-old red wattle piglet that had died suddenly without evidence of significant intercurrent disease. Only one nonred wattle pig, a Yorkshire-Landrace cross, was diagnosed with cardiac rhabdomyoma during this time period. This four-month-old pig had died suddenly without significant intercurrent disease.

Macroscopic lesions were found in two of the stillborn red wattle-cross piglets. Multiple tan, firm, discrete nodules, 0.5 mm–1.0 mm in diameter, were randomly distributed throughout the ventricular myocardium, including the interventricular septum. Formalin-fixed tissues obtained at necropsy were routinely processed, sectioned (6 μ m), and stained with hematoxylin and eosin (H and E). Additional sections were stained with phosphotungstic-acid hematoxylin (PTAH) and periodic acid-Schiff (PAS) with and without diastase.

Histologically, multiple myocardial nodules were present in eight of the nine red wattle and red wattle-cross piglets comprising the three consignments. The variably sized (500 μ m–1 mm), circumscribed but nonencapsulated, foci consisted of interwoven bundles and fascicles

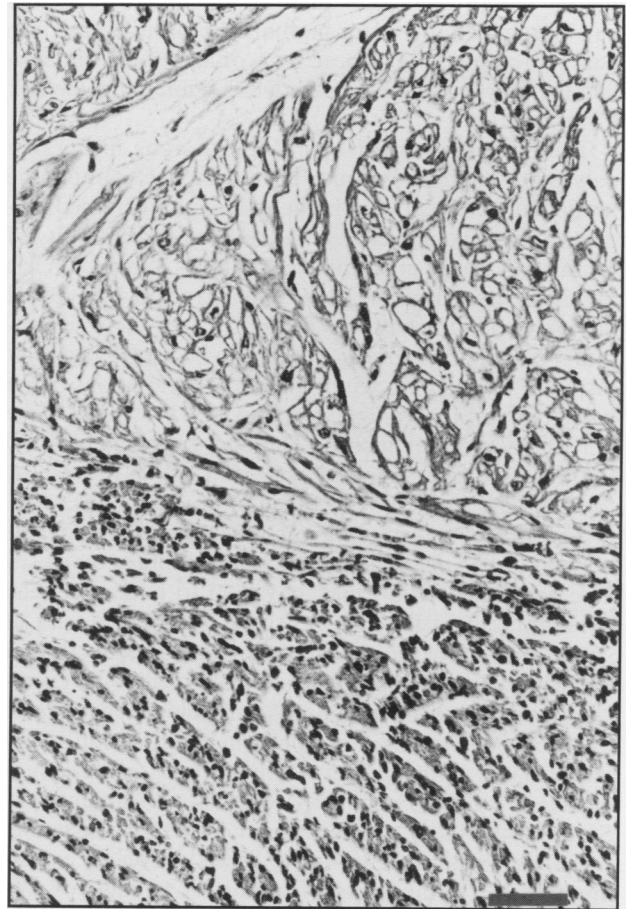


Figure 1. Large vacuolated cells of the circumscribed cardiac rhabdomyoma (top) contrast with the smaller, compact, normal cardiac myofibers (bottom). (Hematoxylin and eosin; bar represents 70 μ m).

of cells cut in different planes. Continuity with adjacent myofibers was evident in a few of the nodules. Large, clear, perinuclear vacuoles, bordered by eosinophilic flocculent cytoplasm, were present in all cells, but they were most obvious in those cut in cross section. Fine, diastase-labile, PAS-positive granules confirmed the presence of intracytoplasmic glycogen within many of the cells. Occasionally, fine, fibrillar, eosinophilic strands attached the nuclear envelope to the peripheral cytoplasm, which is typical of the "spider-cells" of cardiac rhabdomyomas (1,4). Cytoplasmic cross striations similar to those of normal myofibers were visible periodically with H and E staining but were accentuated by staining with PTAH. Although an occasional binucleate cell was observed, most cells contained single, oval, 7 μ m \times 12 μ m, nuclei with finely stippled chromatin. There was mild anisokaryosis, but mitoses were not present.

Cardiac rhabdomyomas of red wattle and red wattle-cross piglets are macroscopically and histologically similar to those previously reported in domestic animals (1,5,6) and humans (4,7). Although they bear some histological resemblance to Purkinje cells, cardiac rhabdomyoma cells share ultrastructural features of both

cardiac myofibers and Purkinje cells (4,6), creating uncertainty as to their histogenesis. The relative paucity of poorly oriented myofibrils, abundant glycogen, binucleation, and desmosomal intercellular junctions in rhabdomyomas are characteristics of Purkinje cells (4,8), but intercalated discs, which are exclusive to cardiac myofibers, are also present in some rhabdomyoma cells (4,8). This combination of ultrastructural features has led to the hypotheses that cardiac rhabdomyomas arise from either two types of fibers (4) or a pluripotential embryonic cell (1).

Not only is the histogenesis of cardiac rhabdomyomas unresolved, but even the nature of the lesion is disputed. It has been suggested that they are true neoplasms, hamartomas, lesions of anomalous glycogen storage, or gigantism of myocardial fibers (1,4). Current dogma favors cardiac rhabdomyomas as hamartomas rather than true neoplasms (7), but even this is debatable, as a hamartoma is defined as a focal excessive overgrowth of mature normal cells indigenous to the tissue (9).

The occurrence of cardiac rhabdomyomas in still-born and neonatal red wattle piglets in the absence of heart failure concurs with previous reports of the congenital and incidental nature of these lesions in pigs (1,2,5,6). However, the absence of intercurrent disease in one red wattle piglet suggests that cardiac rhabdomyomas may potentially cause sudden death, perhaps by interfering with normal myocardial conduction, as occurs in people (7).

In humans, pediatric cardiac rhabdomyomas are frequently associated with tuberous sclerosis, a hereditary, autosomal-dominant syndrome characterized by multisystemic anomalies (10), but they are occasionally detected in otherwise normal infants (4). The overwhelming prevalence of cardiac rhabdomyomas in 8 of 11 red wattle pigs submitted to VLSB, compared to 1 of 4999 pigs of other breeds, evinces a breed predisposition towards these lesions. Obviously, pedigree analysis and heritability studies are necessary to further define the transmission and heritability of cardiac rhabdomyomas in red wattle and red wattle-cross pigs.

Acknowledgments

I thank Drs. Grant Maxie, Gary Thomson, Nigel Palmer and Murray Hazlett for their assistance, and Helga Hunter, Department of Pathology, Ontario Veterinary College, for translating German articles and for her technical expertise.

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